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Case Report: We present a case of isolated plexiform neurofibroma of vulva which presented as white lesion of vulva. A tissue biopsy was done and the histopathological analysis-demonstrated plexiform neurofibroma with spindle shaped cells with wavy nuclei and bland nuclear chromatin. The immunohistochemical staining showed strong positivity for S-100 protein which has a strong correlation with neurofibromatosis-1. No systemic sign of neurofibromatosis was found.

Conclusion: Plexiform neurofibroma may present as isolated white lesion of the vulva without any systemic involvement in the form of neurofibromatosis. So a tissue biopsy is mandatory for definitive diagnosis of any white lesion of vulva.

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CASE REPORT

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Keywords: Vulva, White lesion, Plexiform neurofibroma

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INTRODUCTION

Neurofibroma is one of the uncommonly occurring mesenchymal lesions in vulvo-vaginal region. Neurofibroma as such is a disease of the peripheral nervous system and occurs most commonly in the extremities. Neurofibromas have been categorized as cutaneous neurofibromas (both localized and diffuse types), intraneural neurofibromas (localized and plexiform), massive soft tissue neurofibromas (solitary or multiple) and sporadic neurofibromas or those associated with neurofibromatosis-1 (NF-1) [1, 2]. Plexiform neurofibroma is usually recognized as a pathognomonic criterion of NF-1 (or von Recklinghausen's disease). It may also occur as a solitary lesion arising in a nerve root [3]. These lesions commonly involve labia and clitoris among the female genital tract. Lesions may also affect the vagina, cervix, endometrium myometrium, and ovary and may be associated with urinary tract neurofibromatosis [4]. We present a case of asymptomatic isolated vulvar neurofibroma with unusual presentation as white lesion of vulva without systemic involvement. Already known major differential diagnosis of hypopigmented lesion of vulva are lichen sclerosis, squamous cell hyperplasia, squamous cell carcinoma in situ and Paget's disease of vulva.

CASE REPORT

A 57-year-old female presented in the gynecology outpatient department with complains of postmenopausal

discharge per vaginam, irregular bleeding per vaginam and post coital bleeding since last 36 months. She was a multiparous female, a mother of five children and was menopausal for 15 years. She had past history of pulmonary Koch for which she took antitubercular treatment for two years. There was no family history of neurofibromatosis. On genital examination, she was found to have flat hypopigmented vulvar lesions involving labia minora and labia majora (Figure 1). Clitoris was healthy. Rest of the external genitalia looked healthy. The patient was aware of the lesion for past one year but did not seek any medical treatment. On speculum examination cervix was suspicious in appearance, flushed up with vaginal vault. Rest of the vaginal mucosa was healthy. On systemic examination, there were no swelling or café-au-lait spots over the body. No nerve involvement was observed. On ultrasonography uterus measured 7.53x3.99x2.95 cm with normal myometrium and endometrium. Thickness of endometrial cavity containing fluid measured 11.7 mm. Bilateral ovaries were normal. Cervical cytology showed atrophic smear. After a course of antibiotics for cervicitis, colposcopy showed inflammatory changes only and a punch biopsy was taken. A vulvar biopsy from the white lesion of vulva and an endometrial sampling were also taken.

Histopathologically, microscopic examination of vulvar biopsy revealed focally atrophic epidermis. Dermis showed fascicles and bundles. The tissue comprised of spindle shaped cells with wavy nuclei and bland nuclear chromatin. No atypia or necrosis was seen in the tissue. A diagnosis of plexiform neurofibroma was obvious on microscopic examination (Figure 2). Immunohistochemically, the tumor was strongly positive for S-100 protein (Figure 3) which is seen strongly in association with NF-1. Endometrial histopathology showed chronic endometritis and cervical biopsy showed

Figure 2: Microscopic examination of plexiform neurofibroma (H&E stain, x100).

Figure 3: Immunohistochemical staining for S-100 protein was positive in plexiform neurofibroma (S-100, x200).

Figure 1: External genitalia on examination

chronic cervicitis. At third, sixth and twelfth month follow-up patient is healthy with no complains. There are no color changes, no mass lesion in (white lesion) involved area of vulva.

DISCUSSION

Neurofibroma of female genital system can be categorized according to organ involvement into three categories: 1. Vulvar 2. Clitoral 3. Pelvic (cervical, endometrial, myometrial and ovarian). Most of the cases reported to have vulvar, clitoral and pelvic neurofibromas presented as mass lesions. Till date, literature on isolated vulvar neurofibroma is scarce [5]. Among isolated vulvar neurofibromas, the association is found with trauma such as episiotomies or other vulvar injuries [6]. Approximately, half of all vulvar neurofibromas are found in women with neurofibromatosis. Our patient did not have any features of von Recklinghausen's disease.

There are two major concerns in such patients:

- 1) How these patients should be evaluated?
- 2) How these patients should be followed-up?

Our patient had hypopigmented lesion of vulva which was subsequently diagnosed to have plexiform neurofibroma. According to current nomenclature, such type of lesions is being categorized as non-neoplastic epithelial disorders of skin and mucosa (previously leukoplakia, etc). These lesions are associated with dysplasia and cancer. These lesions do not show significant colposcopic findings and hence the investigation of choice should be tissue diagnosis in such patients. The present case did not have any urinary symptoms so cystoscopy was not performed.

After 12 months of regular follow-up, our patient did not develop any new complains. She did not notice any change in color of the lesion, development of mass in external genital region, inguinal lymphadenopathy. We have planned to keep patient on regular follow-up every 2-3 monthly for external genital and pelvic examination.

Till date no such guidelines have been developed for investigations and management of vulvar neurofibromas. As per available literature, data on malignant transformation is also scarce.

CONCLUSION

We can conclude from the available literature that for women having hypopigmented lesions of vulva differential diagnosis of neurofibroma should be kept in mind. Systemic examination for the presence of café-au-lait spots should be performed. Also detailed examination of the genitourinary tract including cystoscopy should also be undertaken in such cases. In cases with no mass lesions the rate of growth of the lesion or change in color should be kept in mind for the possible development of malignancy.

Author Contributions

Sunita Arora – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Rupneet Kaur – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

Poonam Goel – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

RPS Punia – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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